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# CLUSTER HEADACHE

Cluster headache (CH) always involves pain that is one sided (although it can switch sides) and the main defining feature is the association with one or more of the 'cranial autonomic features normally described as follows:

- Reddening and tearing of the eye
- A runny or blocked nostril
- Droopy eyelid
- Constriction of the pupil
- Flushing and facial sweating

Although possible, it is very unusual for these to not occur in cluster headache. These features tend to come and go with each attack, however, some sufferers may continue to experience the constricted pupil and/or a droopy eyelid, especially after frequent attacks.

In most sufferers the headaches often start at the same time of year and at the same time during the day or night. The pain involved is excruciating and is probably one of the most painful conditions known to humans. Female sufferers have described each attack as being more painful than childbirth.

As few as 0.2% (two in a thousand) of the population suffer from CH, approximately the same number as for multiple sclerosis in the UK. Men are more likely to suffer than women, with an estimated male to female ratio of between 5:1 and 7:1, although this ratio appears to be steadily declining. CH can begin at any age, but most sufferers are more likely to start suffering in their 30s or 40s.

It is extremely useful for everyone involved to understand the three standard terms used in CH:

- A **cluster attack** is an individual episode of pain that can last from a few minutes to some hours.
- A **cluster bout** refers to the time period where a number of attacks re-occur, usually lasting some weeks or months.
- A **remission** is the pain-free period between two cluster bouts.

A single attack always focuses on just one side of the head or face although the headache can change sides between attacks and/or bouts. The pain is excruciatingly severe. It is located mainly behind or around the eye, around the top side of the head and within the temple and forehead, although any part of the head or neck can be affected. The headache normally lasts from between 45 minutes and 1½ hours, but can range from between 15 minutes and 3 hours (or more). The pain often reaches its peak very quickly and maintains its intensity over a period of time before ending equally quickly.

There have also been several descriptions by CH sufferers of symptoms similar to those of migraine. Just before an attack starts many sufferers have symptoms such as tiredness and

yawning. Features during an attack can also include nausea, vomiting, and having an over sensitivity to smells, light and sound. However, unlike most migraine sufferers, nine out of ten CH sufferers are usually restless and irritable during an attack, preferring to move around in the hope that this will relieve the pain.

The cluster attack frequency varies from between one every other day to three times a day, however, some sufferers can have up to eight attacks (or more) each day. Regardless of frequency, attacks can often occur rather uncannily at the same time each day.

Alcohol, exercise and increases in environmental temperature can actually trigger an attack. Drinking alcohol can cause an attack within one hour amongst some CH sufferers, unlike migraine sufferers who generally have a headache some hours after a drink. Alcohol does not usually trigger an attack during a period of pain-free remission. The drug nitroglycerine also produces an attack, naturally normally only under clinical conditions. Allergies, sensitivity to food, hormonal changes, and stress do not appear to be triggers of an attack, although this does vary between sufferers.

There are two types of cluster headache, which are classified according to the duration of the cluster bout: Episodic Cluster Headache (ECH) and Chronic Cluster Headache (CCH)**.**

- **Episodic Cluster Headache (ECH)**

  Approximately eight out of ten sufferers of cluster headache have ECH, which is diagnosed when they have a series of bouts, each one lasting more than a week and separated by pain-free remission lasting more than two weeks. Most ECH sufferers have one or two cluster bouts per year, each lasting between 1-3 months. Often, these bouts tend to start during the same month[s] of the year. Although the duration of the cluster bout and the period of remission vary between individuals, these periods tend to remain relatively consistent within the same individual.

- **Chronic Cluster Headache (CCH)**

  The remaining 20% of CH sufferers have CCH, whereby no pain-free remission occurs within one year, or the remission periods last less than four weeks.

There is only a small amount of literature available regarding the long-term outcome of CH, but all the available evidence suggests that it is a lifelong disorder amongst the majority of sufferers.

In one study, about one in ten ECH sufferers became CCH sufferers whereas a third of CCH sufferers became ECH sufferers. A very encouraging piece of information for CH sufferers is that many of them can expect to develop longer periods of remission as they get older.

There is, as yet, no known cause for cluster headache. However, cluster headache has two major clinical features: the trigeminal (nerve) distribution of pain and the associated ipsilateral (same-sided) autonomic symptoms. Firstly, the pain producing innervation (stimulation) of the cranium projects through branches of the trigeminal and upper cervical nerves to the trigeminocervical complex from where nociceptive pathways project to higher centres. Secondly, the accompanying ipsilateral (same-sided) autonomic symptoms suggest cranial

parasympathetic activation (that's the teary eye, runny and blocked nose) and sympathetic hypofunction (the droopy eye and constriction of the pupil).

However, the third and possibly most important clinical feature is the uncanny timing of both attacks and bouts themselves, which originally suggested an involvement in the brain's master-clock: the hypothalamus or more particularly the suprachiasmatic nucleus (SCN). It is now thought that an abnormality within the hypothalamus is the root cause of the pain, which, when in cycle, releases hormones and chemicals that innervate the trigeminal ganglion, in turn causing the domino effect of pain and cranial autonomic symptoms through the trigeminal nerve down one side of the face and head (and sometimes the neck). This theory is backed up by the regularity of attacks (circannual [time of year] and circadian [time of day]), much lower levels of plasma testosterone (in males) during attacks and bouts, and alterations in the natural production of a variety of hormones/chemicals that affect the biologic clock.

Furthermore, PET (positron emission tomography) studies conducted on the brains' of CH sufferers in the late 1990s demonstrated that there is also ipsilateral (same-sided) hypothalamic activation within the brain: there are direct hypothalamic-trigeminal connections and the hypothalamus is known to have a modulatory role on the nociceptive and autonomic pathways. In summary these studies showed an increase in functional activity of the hypothalamus amongst CH sufferers which is not seen in migraine, and is the prime reason why CH is thought to be caused by an abnormality within the hypothalamus. These abnormalities were seen both when sufferers were undergoing an attack and also whilst pain free and were interpreted as an excessive growth of grey cells within the hypothalamus, or more possibly, within the SCN.

There is also evidence that genetic factors may play an important role in CH, and although the type and number of genes involved is unclear, one recent study focussed upon orexin (or hypocretin), a neuropeptide used by the hypothalamus for signaling.

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